26 Systolic Murmurs

MARTIN A. ALPERT

Definition

A *murmur* is a series of vibrations of variable duration, audible with a stethoscope at the chest wall, that emanates from the heart or great vessels. A *systolic murmur* is a murmur that begins during or after the first heart sound and ends before or during the second heart sound.

Technique

Auscultation of the heart and great vessels should take place in a warm, quiet room with the patient's chest exposed. The clinician should use a stethoscope with plastic or rubber tubing 25 to 30 cm (10 to 12 in) long. The stethoscope should be equipped with a stiff diaphragm and a shallow bell. The patient should be examined in the recumbent, sitting, and left lateral decubitus positions. The principal areas of interest are the primary aortic area (second and third intercostal space at the left sternal border), the tricuspid area (fourth intercostal space at the left sternal border), and the mitral area (cardiac apex). Levine and Harvey have recommended that auscultation begin at the cardiac apex, then proceed along the left sternal border from the tricuspid area to the pulmonic area and finally to the aortic area. The clinician should also auscultate the right parasternal region, the right and left base of the neck, the right and left carotid arteries, the left axilla, and the interscapular area. These are areas to which systolic heart murmurs may radiate or from which extracardiac sounds simulating systolic heart murmurs may emanate. The clinician should alternate use of the diaphragm and bell at each location. Before attempting to detect and characterize a systolic murmur, the clinician should define the first and second heart sounds in order to locate systole accurately.

Certain clinical features common to all murmurs must be defined in the patient with a systolic murmur. These characteristics are intensity (loudness), frequency (pitch), quality, duration, configuration, primary location (point of maximum intensity), and site(s) of radiation. The *intensity* of a heart murmur is most effectively gauged using the system originally proposed by Levine. Grade 1 refers to a murmur so faint that it can be heard only with special effort. A grade 2 murmur is faint, but is immediately audible. Grade 3 refers to a murmur that is moderately loud, and grade 4 to a murmur that is very loud. A grade 5 murmur is extremely loud and is audible with one edge of the stethoscope touching the chest wall. A grade 6 murmur is so loud that it is audible with the stethoscope just removed from contact with the chest wall. In general, murmurs with an intensity of grade 4 or higher are accompanied by a palpable thrill. *Frequency* or *pitch* relates to the velocity of blood at the site of origin of the murmur and is designated as high, medium, or low. In general, the higher the velocity, the higher the pitch of the murmur. Blood flow from a high-pressure chamber to a chamber with lower pressure possesses high velocity; hence the associated murmurs are high pitched. Murmurs that emanate from areas of stenosis where velocity is lower are typically low to medium pitched. *Quality* refers to the tonal effect of the murmurs. Frequently used descriptors are blowing, musical, squeaking, whooping, honking, harsh, rasping, grunting, and rumbling. *Duration* refers to the portion of the cardiac cycle that the murmur occupies. Murmurs may be systolic, diastolic, or continuous. Systolic murmurs may be early systolic, midsystolic, late systolic, or holosystolic. Early systolic murmurs begin with the first heart sound and extend to middle or late systole. Midsystolic murmurs begin following a murmur-free interval in early systole and end with a murmur-free interval (of variable duration) in late systole. Late systolic murmurs begin during the last half of systole and may or may not extend to the second heart sound. Holosystolic murmurs begin with the first heart sound and extend to or through the second heart sound. The *configuration* of a murmur refers to its shape. To a large degree it is a function of intensity and duration. Crescendo murmurs progressively increase in intensity. Decrescendo murmurs progressively decrease in intensity. With crescendo–decrescendo murmurs (diamond or kite-shaped murmurs), a progressive increase in intensity is followed by a progressive decrease in intensity. Plateau murmurs maintain a relatively constant intensity. *Location* refers to the point on the precordium where the murmur is heard with maximum intensity. Many systolic murmurs are audible over multiple areas of the precordium. Localizing their point of maximum intensity may aid greatly in determining their site of evolution. Not all heart murmurs radiate. Defining the sites of *radiation* for those that do is important in determining the underlying cause of the murmur.

A variety of physiologic maneuvers and pharmacologic interventions that alter cardiovascular hemodynamics can be used to aid in the characterization and differentiation of cardiac murmurs. The physiologic maneuvers are breathing, standing, sudden squatting, isometric hand grip exercise, Valsalva maneuver, passive leg raising, and attention to the beat following a postextrasystolic pause. Breathing produces a greater effect on the right side of the heart than the left side. Inspiration increases venous return to the right side of the heart by increasing flow in the vena cava, but decreases venous return to the left side of the heart due to pooling of blood in pulmonary venous capacitance vessels. The effects of inspiration on systolic murmurs can be accentuated by employing Mueller's maneuver (forced inspiration on a closed glottis). Expiration decreases venous return to the right side of the heart by reducing vena cava flow, but increases venous return to the left side of the heart due to collapse of pulmonary venous capacitance vessels. Sudden standing from a sitting or recumbent position decreases venous return, first to the right side of the heart and then to the left side of the heart. This results in a decrease in

stroke volume from those chambers. Recumbency increases venous return first to the right and then to the left side of the heart. Squatting produces a simultaneous increase in venous return first to the right and then to the left side of the heart, and an increase in peripheral vascular resistance. This results in an increase in systemic blood pressure and in stroke volume (occasionally accompanied by a reflex bradycardia). Isometric hand grip exercise for 20 to 30 seconds produces an increase in peripheral vascular resistance, systemic blood pressure, heart rate, cardiac output, left ventricular volume, and left ventricular filling pressure. Valsalva's maneuver consists of four distinct phases. During the initial phase (phase 1), there is a transient increase in left ventricular output due to an increase in intrathoracic pressure. During the strain phase (phase 2), there is a decrease in venous return, first to the right and then to the left side of the heart. Heart rate increases, but stroke volume, mean arterial pressure, and pulse pressure all decrease. During the release phase (phase 3), venous return begins to increase. During the overshoot phase (phase 4), venous filling of the right and left ventricles and heart rate return to normal or are slightly increased. Left-sided events generally lag behind right-sided events by 6 to 8 beats. Passive leg raising increases venous return, first to the right and then to the left side of the heart. The pause following an extrasystole (usually a ventricular premature beat) permits increased ventricular filling, thus enhancing myocardial contractility. The effects of these physiological maneuvers on individual systolic murmurs are discussed in the section entitled Clinical Significance.

The pharmacologic interventions used most commonly in clinical practice are amyl nitrite administration and intravenous infusion of alpha-adrenergic agonists (phenylephrine or methoxamine). Inhalation of amyl nitrite for 10 to 15 seconds produces a marked decrease in peripheral vascular resistance followed by an increase in stroke volume and venous return. Phenylephrine, administered intravenously in a dosage of 0.5 mg, elevates systolic pressure approximately 30 mm Hg for 3 to 5 minutes. Methoxamine, administered intravenously in a dosage of 3 to 5 mg, elevates systolic pressure 20 to 40 mm Hg for 10 to 20 minutes. Both drugs may also produce reflex bradycardia and decrease myocardial contractility. The effects of these pharmacologic interventions on individual systolic murmurs are discussed in the section entitled Clinical Significance.

Systolic murmurs should not be considered in isolation. Quite frequently, other elements of the cardiovascular examination prove to be as important or more important in identifying the cause of the murmur. The best example of this is an ostium secundum atrial septal defect. Here, the systolic murmur is a nondescript pulmonic flow murmur, but the diagnosis is secured by detecting fixed and wide splitting of the second heart sound. Attention to other elements of the cardiovascular examination may also provide important clues to the severity of the abnormality causing the murmur.

Basic Science

Murmurs are created by disturbance of laminar blood flow (i.e., turbulence), but turbulence per se does not produce adequate acoustic force to be audible at the chest wall. The most widely accepted theory concerning the generation of murmurs was popularized by Bruns and incorporates the concept of *vortex shedding*. Vortices are tiny eddies created by an obstruction to the laminar flow of blood. The concept of vortex shedding can be simplified by employing a familiar analogy—a boulder protruding through the surface of a fast-moving stream. The undisturbed water flows without interruption until it hits the boulder. The boulder causes the stream to separate and generate vortices, or tiny eddies that move in a spiral fashion and are shed in the general direction of the flow of the stream. As the vortices are shed, they leave in their place wakes, which are areas of relatively "still water." Water rapidly moves in to fill the wakes left by vortex shedding. The sound that one hears when water is rushing around the boulder is generated by the filling of wakes left by the shedding of vortices.

A similar situation exists in the cardiovascular system. Deformity of valvular structures, valvular stenosis, and discontinuity in a wall of the heart or in the great vessels may provide a site for vortex shedding. Moreover, substantial vortex shedding can result from increased flow over the normal protrusions and irregularities in the heart or great vessels. The response of blood moving to fill the wakes left by the shedding of vortices is an efficient mechanism for the generation of sound and is capable of giving rise to sustained vibrations that are audible at the chest wall (i.e., murmurs).

An important variable in this theory is *velocity*. At normal velocities in the cardiovascular system, vortex shedding is minimal and the flow of blood is not audible. When the velocity of blood flow increases substantially (as in high cardiac output states), vortex shedding increases to the point where the frequencies generated are audible to the human ear (with the help of a stethoscope). When additional areas of disruption of laminar flow are present, vortex shedding is increased even at normal velocities, and murmurs are generated. Where one listens in relation to where the vortices are being shed is important in terms of the pitch of the murmur. As vortices are shed around an obstruction, some of them coalesce downstream to form a lower frequency than that present at the site of shedding. Thus, the sounds generated upstream are generally a more accurate reflection of the true shedding frequency than those heard downstream where vortices have had the opportunity to coalesce.

Clinical Significance

Systolic murmurs may be classified as ejection murmurs, regurgitant murmurs, or extracardiac sounds that simulate systolic heart murmurs. *Ejection murmurs* emanate from the semilunar valves or surrounding structures (i.e., the aortic or pulmonic root). *Regurgitant murmurs* are created when blood flows from a high-pressure "donor" chamber to a low-pressure "recipient" chamber. Table 26.1 is a summary of systolic ejection and regurgitant heart murmurs and extracardiac sounds. Figure 26.1 shows selected characteristics of the most common of these murmurs and their relationship to the heart sounds.

Systolic murmurs may be further subclassified as functional or organic. *Functional systolic murmurs* occur in the absence of cardiac structural abnormalities. They are frequently encountered in healthy individuals, but may also accompany a variety of high cardiac output states. *Organic systolic murmurs* evolve from structural abnormalities in the heart or great vessels. Systolic ejection murmurs may be functional or organic, but systolic regurgitant murmurs indicate organic heart disease.

Table 26.1
Differential Diagnosis of Systolic Murmurs

I. Ejection murmurs
  A. Functional
    1. Still's murmur and its adult variant
    2. Flow murmur emanating from the root of the pulmonary artery
    3. Murmur associated with high cardiac output states
    4. Flow murmurs associated with aortic or pulmonary valvular insufficiency
  B. Organic
    1. Valvular aortic stenosis
    2. Aortic sclerosis
    3. Discrete subvalvular aortic stenosis (web or tunnel)
    4. Supravalvular aortic stenosis
    5. Hypertrophic obstructive cardiomyopathy
    6. Pulmonary valvular stenosis
    7. Pulmonary infundibular stenosis
    8. Atrial septal defect
    9. Tetralogy of Fallot
II. Regurgitant murmurs
  A. Functional: none
  B. Organic
    1. Mitral regurgitation
      a. Rheumatic
      b. Papillary muscle dysfunction
      c. Mitral valve prolapse
      d. Acute
    2. Tricuspid regurgitation
      a. Chronic
      b. Acute
    3. Ventricular septal defect
      a. Roger's type (small and large)
        (1) Without pulmonary hypertension
        (2) With pulmonary hypertension
      b. Slitlike
III. Extracardiac sounds simulating systolic heart murmurs
  A. Subclavian (supraclavicular/brachiocephalic) murmur
  B. Internal mammary souffle
  C. Carotid artery bruits
  D. Coarctation of the aorta
  E. Murmurs emanating from a dilated aortic or pulmonary artery root
  F. Patent ductus arteriosus with pulmonary hypertension

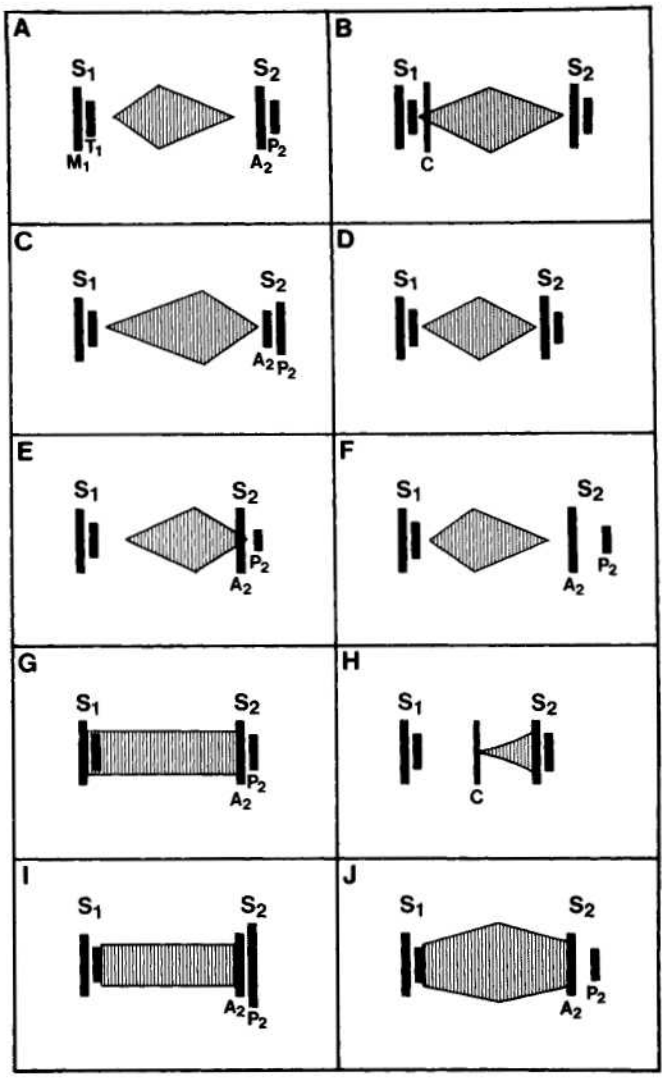

Figure 26.1
Selected characteristics of common systolic heart murmurs. Abbreviations: S_1 = first heart sound, S_2 = second heart sound, M_1 = mitral component, T_1 = tricuspid component, A_2 = aortic component, P_2 = pulmonic component, C = click. A: functional systolic ejection murmur; note early peaking. B: mild aortic valvular stenosis; note relatively early peaking and systolic ejection click. C: severe aortic stenosis; note late peaking and decreased intensity of A_2. D: hypertrophic obstructive cardiomyopathy. E: severe pulmonary valvular stenosis; note late peaking with murmur extending through A_2 and delayed appearance of P_2. F: atrial septal defect; note wide splitting of S_2. G: uncomplicated mitral regurgitation; note holosystolic murmur extending through A_2. H: mitral valve prolapse; note late systolic murmur ushered in by midsystolic click. I: tricuspid regurgitation due to pulmonary hypertension; note holosystolic murmur beginning with T_1 and early, loud P_2. J: uncomplicated ventricular septal defect; note loud, holosystolic murmur with midsystolic accentuation and slightly delayed P_2.

Ejection Murmurs

Functional systolic ejection murmurs include pulmonic flow murmurs in patients with either normal or increased pulmonary artery or aortic flow. The most common functional systolic ejection murmur in adults is probably a variant of Still's murmur, the so-called *innocent murmur of childhood.* It is a short, buzzing, pure, medium-pitched, nonradiating, midsystolic murmur heard best along the upper left sternal border. It is thought to result from vibrations set in motion by the pulmonic valve. A less frequently encountered functional pulmonic flow murmur, occurring predominantly in children and adolescents, emanates from the root of the pulmonary artery. It is midsystolic and similar in location, but less uniform than Still's murmur. It is high pitched, often blowing in nature, and is similar to the flow murmur heard in patients with the "straight-back syndrome." Functional systolic ejection murmurs may also result from hyperdynamic blood flow over a normal pulmonic or aortic valve. Such murmurs are commonly associated with high cardiac output states such as thyrotoxicosis, anemia, infection, fever, arteriovenous fistula, beriberi, the hyperkinetic heart syndrome, or pregnancy. They may also be audible after exercise or with anxiety.

Systolic flow murmurs associated with *aortic or pulmonic valvular insufficiency* may also be included in this category. These murmurs are typically midsystolic. They begin in early systole, peak in early to midsystole, end well before the onset of the second heart sound, have variable intensity, and do not radiate. These murmurs are usually medium pitched and have a crescendo–decrescendo configuration. Although the vibrations that produce these murmurs em-

anate from both semilumar valves, the murmurs are best (often exclusively) heard at the pulmonic area of the precordium because of the proximity of the pulmonic valve to the chest wall.

The intensity of functional systolic ejection murmurs varies, but generally ranges from grades 1 to 3. The intensity typically increases during phase 3 (release phase) of the Valsalva maneuver, following a post-extrasystolic pause and after inhalation of amyl nitrite. The intensity may decrease with isometric hand grip exercise or intravenous administration of alpha-adrenergic agonists.

Organic systolic ejection murmurs include those associated with valvular aortic stenosis, aortic sclerosis, supravalvular aortic stenosis, subvalvular aortic stenosis, hypertrophic obstructive cardiomyopathy, valvular pulmonic stenosis, pulmonary infundibular stenosis, atrial septal defect, and tetralogy of Fallot.

The murmur of *valvular aortic stenosis* typically has a crescendo–decrescendo configuration and a low to medium pitch. The murmur is commonly described as harsh, rasping, grunting or rough. Best heard over the primary and secondary aortic areas, the murmur is transmitted widely over the precordium and radiates to the carotid arteries. The intensity of the murmur is variable (usually grade 2, 3, or 4), and increases with passive leg raising, sudden squatting, 5 or 6 beats into phase 3 of the Valsalva maneuver (release phase), following a post-extrasystolic pause and after amyl nitrite administration. The murmur tends to fade during phase 2 of the Valsalva maneuver (strain phase), with isometric hand grip exercise and occasionally with intravenous administration of alpha-adrenergic agonists. In elderly persons the murmur may be more intense and high pitched over the mitral area. It commonly takes on a musical or wheezing quality and may be confused with mitral valve regurgitation. The crescendo–decrescendo configuration helps to differentiate aortic stenosis from mitral valve regurgitation. The murmur may peak in either mid or late systole. There is a tendency for the murmur to peak progressively later in systole as stenosis become more severe. Other signs that suggest severity of valvular aortic stenosis include a delayed carotid upstroke (the single best criterion on physical examination), diminished intensity or paradoxical splitting of the second heart sound, and left ventricular hypertrophy. The presence of a systolic ejection click suggests that valve mobility is reasonably well preserved; it mitigates against severe stenosis.

Aortic sclerosis results from degeneration and calcification of the aortic cusps, predominantly at their base. This abnormality produces a murmur that is identical in character to aortic valvular stenosis but does not result in a pressure gradient over the aortic valve. The murmur typically peaks in midsystole and is accompanied by a normal second heart sound and carotid pulse upstroke. The responses to physiologic and pharmacologic interventions are identical to those of functional systolic ejection murmurs. Echocardiographic studies suggest that aortic sclerosis is common in the elderly and may be the single most common cause of systolic murmur in this population.

Discrete subaortic stenosis, resulting from a fibrous web or fibromuscular tunnel, produces a murmur that is indistinguishable from aortic stenosis. An ejection click is typically absent. The absence of calcium and poststenotic dilatation of the aorta on chest x-ray or fluoroscopic examination of a patient with apparently "severe aortic stenosis" suggests the diagnosis.

Supravalvular aortic stenosis also produces a murmur indistinguishable from valvular aortic stenosis. The murmur is typically most intense over the primary aortic area and radiates to both carotid arteries, but the amplitude of the carotid and brachial pulses may be unequal (greater on the right side) due to the orientation of the jet of blood that traverses the area of stenosis. An ejection click is typically absent. Supravalvular aortic stenosis usually becomes apparent early in childhood, and may be associated with mental retardation and elfin facies.

The systolic murmur associated with *hypertrophic obstructive cardiomyopathy (HOCM),* also known as idiopathic hypertrophic subaortic stenosis, results from dynamic left ventricular outflow tract obstruction. Left ventricular outflow obstruction is probably related to systolic anterior motion of the anterior mitral valve leaflet caused by a Venturi (suction) effect in the outflow tract resulting from rapid ejection of blood. Recent evidence suggests that concurrent mitral valve regurgitation may contribute to the murmur in some cases. The murmur of HOCM is typically medium pitched, has a crescendo–decrescendo configuration, and is heard best along the left sternal border. There is radiation to the base of the neck, but not into the carotid arteries. The murmur is similar in quality to that of aortic valve stenosis, but is somewhat less harsh and slightly higher pitched. The intensity of the murmur is quite variable. The murmur of HOCM becomes louder during phase 2 of Valsalva's maneuver (strain phase), with standing, after a post-extrasystolic pause, and following amyl nitrite inhalation. All of these maneuvers or interventions reduce left ventricular volume, increasing the subaortic pressure gradient. The murmur decreases in intensity with recumbency, passive leg raising, squatting, isometric hand grip, or following intravenous administration of alpha-adrenergic agonists. The murmur of HOCM is commonly accompanied by a fourth heart sound, a double or triple apical impulse, and a bisferiens (spike and dome) carotid pulse. A thrill is occasionally palpable over the left sternal border.

Valvular pulmonic stenosis is usually congenital in origin. Severe cases are apt to be encountered more in childhood than adulthood. The murmur of valvular pulmonic stenosis is midsystolic (with respect to right heart events). It begins well after the first heart sound and ends before the pulmonic component of the second heart sound (which may be delayed in severe cases), but often extend through the aortic component of the second heart sound. The murmur has a crescendo–decrescendo configuration. There is a strong tendency for the murmur to peak later in systole as stenosis becomes more severe. The murmur is best heard over the second intercostal space at the left sternal border and does not radiate. Its quality is similar to that of valvular aortic stenosis. The intensity of the murmur is variable but is generally grade 3 or higher; it increases during phase 3 of the Valsalva maneuver (release phase), with inhalation of amyl nitrite, and occasionally with passive leg raising and following a post-extrasystolic pause. The murmur is commonly accompanied by a right ventricular lift and thrill palpable along the upper left sternal border. A fourth heart sound (right sided) may be audible at the lower left sternal border. A pulmonic ejection click is frequently audible (during expiration) over the pulmonic area except in very severe cases. Patients with congenital pulmonary valvular stenosis commonly, but not invariably, have moon facies and hyperteleorism.

Infundibular pulmonic stenosis is present in approximately

10% of cases of right ventricular outflow obstruction. It is usually accompanied by a ventricular septal defect, but may also occur as an isolated abnormality. The murmur of isolated infundibular pulmonic stenosis is identical to that of valvular pulmonic stenosis except that it is best heard lower along the left sternal border (third intercostal space) and is rarely accompanied by an ejection click.

The murmur associated with *atrial septal defect* is caused by increased blood flow in the right ventricular outflow tract. Accordingly, it is identical in character to the functional pulmonic ejection murmurs previously described. The presence of fixed and wide splitting of the second heart sound, not the nonspecific pulmonic flow murmur, confirms the presence of atrial septal defect.

The murmur associated with *tetralogy of Fallot* (ventricular septal defect, infundibular pulmonic stenosis, right ventricular hypertrophy, and overriding aorta) emanates predominantly from the ventricular septal defect when pulmonic stenosis is mild, but from the pulmonary outflow tract when pulmonic stenosis is moderate to severe. Tetralogy of Fallot with mild pulmonic stenosis is characterized by a loud (usually grade 4 or higher), harsh, midsystolic, crescendo–decrescendo murmur. The murmur is widely audible over the precordium and peaks in late midsystole. With moderate to severe pulmonic stenosis, the murmur peaks progressively earlier in systole as much of the blood flow is shunted through the ventricular septal defect into the aorta. In extreme cases a short early systolic aortic flow murmur may be all that is audible. Tetralogy associated with mild pulmonic stenosis may be accompanied by a delayed and diminished pulmonic component of the second heart sound. The pulmonic component is not audible when pulmonic stenosis is moderate to severe. When extreme pulmonic stenosis is present, an aortic ejection click may be audible.

Regurgitant Murmurs

Systolic regurgitant murmurs include the many variations of mitral valve regurgitation, tricuspid valve regurgitation, and ventricular septal defect. Depending on its etiology and pathogenesis, mitral regurgitation may produce four discrete auscultatory patterns. Classic mitral valve regurgitation is typified by the systolic murmur of a rheumatic mitral valve. Other variations include mitral regurgitation associated with papillary muscle dysfunction, acute mitral regurgitation, and mitral valve prolapse.

The murmur of *rheumatic mitral valve regurgitation* is high pitched, blowing, and best heard at the cardiac apex with radiation to the axilla. It is holosystolic, starting with the first heart sound and extending to and sometimes through the aortic component of the second heart sound. Typically plateau in configuration, the murmur occasionally has late systolic accentuation. The intensity of classic mitral valve regurgitation is quite variable. The intensity increases with squatting, isometric hand grip exercise, and intravenous administration of alpha-adrenergic agonists. It typically decreases with amyl nitrite inhalation. As a result of the volume overload state associated with valvular mitral regurgitation, a third heart sound is commonly audible at the apex. Dilated cardiomyopathy can produce a mitral regurgitation murmur similar in character to rheumatic mitral valve regurgitation.

Mitral regurgitation associated with papillary muscle dysfunction is typically a midsystolic, crescendo–decrescendo, nonradiating murmur localized to the lower left sternal border. The murmur is medium to high pitched and commonly has a musical or cooing quality. The intensity is generally less than grade 4. Mitral regurgitation murmurs associated with left ventricular dilatation (e.g., from dilated cardiomyopathy) may occasionally present in this fashion.

Mitral valve prolapse (idiopathic or secondary) is a common cause of mitral regurgitation. Classically, mitral valve prolapse is characterized by a medium-pitched late systolic murmur ushered in by a midsystolic click. The murmur is best heard over the apex and generally does not radiate. The murmur commonly, but not invariably, has a crescendo configuration as it reaches the aortic component of the second heart sound. The intensity is quite variable. The intensity increases and the murmur starts earlier in systole with maneuvers or interventions that decrease left ventricular volume (and therefore increase the severity of prolapse), for example, standing, phase 2 (strain phase) of the Valsalva maneuver, and following amyl nitrite inhalation. These interventions also cause the midsystolic click to migrate closer to the first heart sound. Maneuvers and interventions that increase left ventricular volume decrease the intensity of the murmur. These include passive leg raising, recumbency, a post-extrasystolic pause, squatting, isometric hand grip exercise, and intravenous administration of alpha-adrenergic agonists. Such maneuvers and interventions cause the click to return to its original location in midsystole. Not infrequently, the murmur of mitral valve prolapse is holosystolic (either spontaneously or following interventions to decrease left ventricular volume). Such a murmur generally retains a medium pitch, but may radiate to the axilla. It may reach an intensity of grade 5 or 6 and may have a musical whooping or honking quality.

Acute mitral valve regurgitation is most commonly caused by chordae tendineae rupture, but may also result from papillary muscle rupture, infective endocarditis, or trauma. The murmur of acute mitral regurgitation is typically decrescendo and of variable intensity (usually grade 3 or higher). It begins with the first heart sound and decreases in intensity throughout systole, occasionally terminating before the aortic component of the second heart sound. Best heard at the cardiac apex, the murmur typically radiates to the axilla and may be audible along the cervical spine or at the top of the head in selected cases. The murmur is lower pitched than that of rheumatic mitral regurgitation, often possessing a harsh quality reminiscent of valvular aortic stenosis. A third and fourth heart sound may be audible at the apex.

The murmur of *tricuspid valve regurgitation* is typically a high-pitched, blowing, holosystolic, plateau, nonradiating murmur best heard at the lower left sternal border. The intensity is variable, but tends to increase during inspiration (Carvallo's sign), with passive leg raising, after a post-extrasystole pause, and following amyl nitrite inhalation. The intensity of the murmur tends to correlate positively with the severity of regurgitation. Right ventricular enlargement may displace the location of the murmur leftward. Right ventricular failure may abolish respiratory variation. When tricuspid regurgitation is caused by pulmonary hypertension, a pulmonic ejection click may be audible. Severe tricuspid regurgitation is commonly accompanied by a third heart sound emanating from the right ventricle and best heard at the lower left sternal border. Severe tricuspid regurgitation typically produces an accentuated jugular "cv" wave and may produce hepatic congestion with a pulsatile liver.

The murmur of acute tricuspid valve regurgitation (due to infective endocarditis or trauma) is similar in character to that previously described, but often possesses a decrescendo configuration (similar to acute mitral regurgitation) and may terminate before the second heart sound.

The auscultatory findings associated with *ventricular septal defect* are variable, depending on a variety of morphologic and hemodynamic considerations. The systolic murmur associated with a Roger's-type ventricular septal defect (regurgitant jet flows directly into the right ventricular outflow tract) in patients with low pulmonary vascular resistance is a low to medium pitched, holosystolic murmur with midsystolic accentuation. The murmur is heard best over the third and fourth intercostal space at the left sternal border but is widely audible over the entire precordium. The intensity of the murmur is typically grade 3 or higher. Amyl nitrite inhalation causes the murmur of an uncomplicated ventricular septal defect to decrease, whereas alpha-adrenergic agonists cause no change or an increase in intensity. A precordial thrill and third heart sound (from left ventricular volume overload) frequently accompany the murmur. The pulmonic component of the second heart sound is occasionally delayed.

If pulmonary hypertension produces equalization or reversal of shunt flow in a Roger's-type ventricular septal defect (Eisenmenger's complex), the systolic murmur emanating from the septal defect may disappear entirely or be replaced by an early-peaking, medium-pitched, midsystolic murmur emanating from a dilated aortic root. Other auscultatory signs of pulmonary hypertension, including a loud pulmonic component of the second heart sound, a pulmonary ejection click, the early diastolic murmur of pulmonary valve insufficiency (Graham-Steel murmur), and right-side Austin-Flint murmur, may accompany the relatively nondescript systolic murmur. Physiologic maneuvers and pharmacologic interventions have little effect on small defects when pulmonary hypertension is present. With large defects, the murmur intensity may increase with amyl nitrite inhalation, but shows no change or decreases following intravenous administration of alpha-adrenergic agonists.

Small, slit-like ventricular septal defects typically produce an early systolic murmur that ends during the early portion of midsystole.

Extracardiac Sounds

Extracardiac sounds that may simulate systolic murmurs include the innocent subclavian murmur, carotid murmurs, the internal mammary souffle, coarctation of the aorta, and murmurs associated with a dilated aortic or pulmonic trunk and patent ductus arteriosus associated with pulmonary hypertension.

The *innocent subclavian murmur* (supraclavicular or brachiocephalic systolic murmur) is detected most frequently in children and adolescents. It has a crescendo–decrescendo configuration, an abrupt onset and brief duration in early to midsystole, and is medium to high pitched. Its origin is the brachiocephalic artery, and it is best heard over that region, but it may be heard with attenuation over the aortic and pulmonic areas of the precordium. The murmur disappears with compression of the brachiocephalic artery or hyperextension of the ipsilateral shoulder, a maneuver accomplished by bringing the elbows well behind the back, causing the shoulder girdle muscles to be taut. This murmur is frequently mistaken for an organic systolic murmur. Consequently some children who have it are unnecessarily kept from athletic or other strenuous physical activities.

A soft, high-pitched, early to midsystolic murmur may occasionally be audible along the sternal borders in pregnant women and appears to emanate from one of the internal mammary arteries. It is termed the *internal mammary souffle* and is of no pathological significance.

Bruits that originate in the carotid arteries may occasionally be mistaken for systolic heart murmurs. They are consistently heard best over the carotid arteries and are heard only with attenuation over the aortic and pulmonic areas.

Coarctation of the aorta typically is associated with a systolic murmur. In 50% of the cases a bicuspid aortic valve is present. When the murmur is not caused by valvular stenosis, it is thought to result from rapid blood flow across the stenotic segment of aorta. The murmur of coarctation is medium to high pitched and peaks rather late in systole. It is heard best over the left interscapular area and faintly if at all over the aortic areas of the precordium. The typical peripheral manifestations of coarctation permit relatively easy detection when a thorough examination is performed.

Systolic murmurs may also emanate from a *dilated aortic or pulmonary trunk*. Most commonly, this occurs in association with hypertension in the respective vascular circuit. The murmur is similar in character to that of aortic sclerosis or mild pulmonic stenosis.

Patent ductus arteriosus produces a continuous murmur in patients with normal pulmonary vascular resistance. As pulmonary vascular resistance increases, the diastolic portion of the murmur attenuates. With equalization of pressures in the systemic and pulmonary circuits, the systolic component of the murmur remains, extends through the second heart sound, and ends in early diastole. When pulmonary hypertension produces a right-to-left shunt, the murmur emanating from the ductus disappears and is replaced by a systolic flow murmur emanating from the root of the pulmonary artery.

The cause of a systolic murmur can accurately be ascertained in most cases from the physical examination. The medical history, resting electrocardiogram, and chest x-ray may provide valuable information concerning the impact of the underlying cardiac abnormality on the patient's cardiopulmonary status, but rarely provide specific information useful in characterizing the systolic murmur. Phonocardiography and recording of pulse tracings may be used to confirm clinical suspicions. In this regard, they are most useful as teaching tools. Echocardiography is well suited to the characterization of systolic heart murmurs. A complete examination, employing M-mode, two-dimensional, and Doppler echocardiographic techniques, can accurately identify the cause of virtually any organic systolic heart murmur (and by exclusion confirm the presence of a functional systolic ejection murmur). Limitations exist in the ability of echocardiography reliably to quantify the severity of most cardiac abnormalities that produce systolic murmurs. Despite its potential usefulness in identifying underlying causes of systolic murmurs, echocardiography is too expensive to use as a screening tool. It should be considered only when the diagnosis is in question following examination by an experienced clinician. Cardiac catheterization is rarely needed to define the cause of a systolic heart murmur, but may be of great value in assessing the severity of the cause of the murmur and determining its impact on the heart and circulation.

References

Bruns D. A general theory of the causes of murmurs in the cardiovascular system. Am J Med 1959;27:360–74.

Craig E. Echophonocardiography. In: Heart disease. A textbook of cardiovascular medicine, 2d ed. Philadelphia: W.B. Saunders, 1984;68–87.

Harvey WP. Innocent versus significant systolic murmurs. Curr Prob Cardiol 1966;1(8):1–55.

Leatham A. Systolic murmurs. Circulation 1958;17:601–11.

Leatham A, Leech GJ, Harvey WP, de Leon AC. Auscultation of the heart. In: Hurst JW, Logue BR, Rackley CE, Schlant RC, Sonnenblick EH, Wallace AG, Wenger NK, eds. The heart. Arteries and veins, 6th ed. New York: McGraw-Hill, 1986.

Levine S. The systolic murmur: its clinical significance. JAMA 1934;101:436–38.

Reddy PS, Shaver JA, Leonard JJ. Cardiac systolic murmurs: pathophysiology and differential diagnosis. Prog Cardiovasc Dis 1971;14:1–37.

Tavel M. Clinical phonocardiography and external pulse recording, 3d ed. Chicago: Year Book Medical Publishers, 1978;124–46, 165–84.